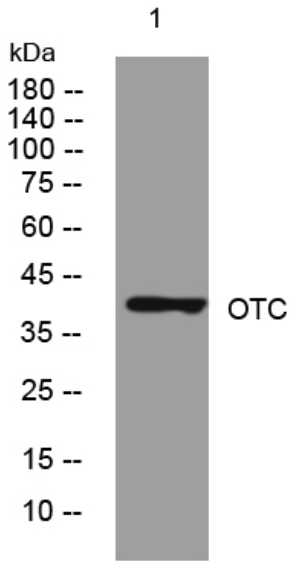
OTC rabbit pAb

Catalog No	BYab-07946
Isotype	IgG
Reactivity	Human; Mouse;Rat
Applications	WB
Gene Name	OTC
Protein Name	OTC
Immunogen	Synthesized peptide derived from human OTC AA range: 275-325
Specificity	This antibody detects endogenous levels of OTC at Human/Mouse/Rat
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.60% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	Ornithine carbamoyltransferase, mitochondrial (EC 2.1.3.3) (Ornithine transcarbamylase) (OTCase)
Observed Band	38kD
Cell Pathway	Mitochondrion matrix .
Tissue Specificity	Mainly expressed in liver and intestinal mucosa.
Function	catalytic activity:Carbamoyl phosphate + L-ornithine = phosphate + L-citrulline.,disease:Defects in OTC are the cause of ornithine carbamoyltransferase deficiency (OTCD) [MIM:311250]. OTCD is an X-linked disorder of the urea cycle which causes a form of hyperammonemia. Mutations with no residual enzyme activity are always expressed in hemizygote males by a very severe neonatal hyperammonemic coma that generally proves to be fatal. Heterozygous females are either asymptomatic or express orotic aciduria spontaneously or after protein intake. The disorder is treatable with supplemental dietary arginine and low protein diet. The arbitrary classification of patients into the "neonatal" group (clinical hyperammonemia in the first few days of life) and "late" onset (clinical presentation after the neonatal period) has been used to

	differentiate severe from mild forms.,online information:OTCase
Background	This nuclear gene encodes a mitochondrial matrix enzyme. Missense, nonsense, and frameshift mutations in this enzyme lead to ornithine transcarbamylase deficiency, which causes hyperammonemia. Since the gene for this enzyme maps close to that for Duchenne muscular dystrophy, it may play a role in that disease also. [provided by RefSeq, Jul 2008],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western blot analysis of lysates from U2OS cells, primary antibody was diluted at 1:1000, 4°over night